Retinoblastoma occurs when abnormal cells in the retina (the light-sensing area at the back of the eye) grow in an uncontrolled way. It usually occurs in young children, and can affect one or both eyes.

Retinoblastoma is more common in children under 3 years of age, although it can occur at any age.

Risk factors

A risk factor is anything that increases a person's chance of developing a certain condition or disease, such as cancer. In adults, lifestyle and environmental factors (such as smoking or exposure to certain chemicals) can be significant risk factors for developing certain types of cancer. In children, very few risk factors have been identified that increase the chance of developing cancer. For most children with cancer, the underlying cause is unknown.

Even if your child has a risk factor, it does not mean they will develop cancer. Many children with a risk factor will never develop cancer, while others with cancer may have had no known risk factors. Even if a child with a risk factor develops cancer, it is usually hard to know how much that risk factor contributed to the development of their disease.

The causes of retinoblastoma are not well understood, but factors associated with a higher chance of developing retinoblastoma include the following.

Family history

Retinoblastomas are due to a faulty *RB1* gene. This faulty gene can run in families, or it may develop for the first time in the child. Most cases of retinoblastoma do not run in families.

Knowledge of your family's history relating to cancer may change over time.

Symptoms

Symptoms of retinoblastoma may include:

- pupil (the black part of the eye in the middle) that appears white instead of red when a light is shone into it (e.g. in a photograph)
- red or painful eye
- larger than usual eyeball
- cloudiness in the iris (the coloured part of the eye) and the pupil

- eyes that seem to be looking in different directions (also called 'lazy eye').

Note about symptoms

Many conditions – including common childhood infections – can cause these symptoms, not just retinoblastoma. If your child has any of these symptoms and you are concerned, talk to your child's doctor.

Diagnosis

Your child will have a number of tests to investigate their symptoms and confirm a diagnosis of retinoblastoma, including:

- medical history and physical examination
- eye examination
- blood tests
- urine tests
- medical imaging, which may include
    - ultrasound of the eye
    - computed tomography (CT) scan
    - magnetic resonance imaging (MRI)
    - bone scan.

These tests are explained in more detail in How is cancer diagnosed?.

Staging

If your child is diagnosed with retinoblastoma, some of the diagnostic tests will also help to stage the tumour. Staging determines where the tumour is, how big it is, which nearby organs are involved and whether the cancer has spread to other parts of the body. This is important to determine the outlook (prognosis) for your child, and to decide on the best options for treatment.

There are different ways to assess the stage or extent of disease. One of the most common ways to describe stages for retinoblastoma is the International Retinoblastoma Staging System:

- Stage 0 – the tumour has not spread beyond the eye and was treated without surgery, and the eye has not been removed.
- Stage I – the tumour has not spread beyond the eye, the eye has been removed, and there are no cancer cells left.
- Stage II – the tumour has not spread beyond the eye, the eye has been removed, but some

cancer cells are left.
- Stage IIIa – cancer has spread to tissues in the eye socket.
- Stage IIIb – cancer has spread to nearby lymph nodes.
- Stage IVa – cancer has spread to the blood but not the brain or spinal cord, and there may be secondary tumours in other parts of the body.
- Stage IVb – cancer has spread to the brain or spinal cord, and possibly other parts of the body.

The American Cancer Society describes other systems for staging retinoblastoma.

Treatment

Treatment and care of children with cancer is usually provided by a team of health professionals called a multidisciplinary team. Members of this team are specialists in children's cancers – they understand the differences between children's cancer and adult cancer, and each team member brings different skills in managing care to meet the needs of both you and your child.

The team will be led by a childhood cancer specialist (paediatric oncologist). Other members of the team depend on the age of your child and their type of disease, and may change over time as your child's needs change. A list of team members who might make up the multidisciplinary team can be found in The treatment team.

Treatment for retinoblastoma depends on the age of your child, the stage of the disease, the biological features of the cancer and other factors identified during diagnosis. Treatment will be tailored to your child's particular situation, and may involve one or more of the following (see How is cancer treated for more detail).

Surgery

Your child may have surgery to remove the entire eye and part of the optic nerve. This procedure is called enucleation. This is done if your child's eye has already been so damaged by the tumour that there is little or no chance they would be able to see, and the tumour is large, has not responded to treatment or has come back after treatment.

During the surgery, an implant is placed in the eye socket to take the place of the eye. This is attached to the eye muscles so it can move, and a life-like layer can be added to the surface to create an artificial eye.

Chemotherapy

Chemotherapy uses anti-cancer medicines to destroy cancer cells. It is often given as a combination of medicines to try to prevent the cancer cells from becoming resistant to just one or two medicines.

Chemotherapy medicines are given together in courses, often over a few days. Once the body has recovered from the side effects, the next course is given. Most children receive multiple courses of chemotherapy.

Retinoblastoma is usually treated with combination chemotherapy. Chemotherapy can be injected into a vein or given as a pill to help shrink the tumour and avoid surgery to remove the eye, or used after another treatment to destroy any remaining cancer cells. Chemotherapy can also be delivered directly to the eye.

Radiation therapy

Radiation therapy (also called radiotherapy) uses high-energy X-rays or other types of radiation to destroy cancer cells or stop them from growing. Your child may have radiation therapy to the eye (external beam radiation) to treat retinoblastoma. This can sometimes be given as 'plaque brachytherapy', where radioactive material is placed on the inside of a small metal disc, and the disc is placed on the outside of the eye near the tumour for a few days.

Radiation therapy can sometimes be used as an alternative to surgery so that your child can still use their eye.

Radiation therapy can have long-term side effects in children. If the potential benefits outweigh the risks and radiation therapy is included in your child’s treatment, special care will be taken to reduce these risks.

Stem cell transplant

Some children may be treated using a stem cell transplant (also known as a bone marrow transplant), in combination with high-dose chemotherapy.

Other treatments

Retinoblastoma may be treated by freezing and destroying the abnormal cells (called cryotherapy), or using heat from a laser beam to destroy the cancer (thermotherapy or photocoagulation).

Support

Diagnosis of cancer in a child is a very difficult time for the child, their family and their friends. You might feel overwhelmed, scared, anxious or angry. These are all normal feelings. It is very important to seek support from family, friends, health professionals or other services to help you, your child and your family cope with cancer.

Talk to your child's treatment team if you are having difficulties coping.

Living with children's cancer has information about physical, emotional and practical issues during and after diagnosis and treatment. There is also a page with helpful links on where to find support.

The Cancer Council in your state or territory can give you general information about cancer, as well as information on resources and support groups in your local area. Call the Cancer Council Helpline from anywhere in Australia for the cost of a local call on **13 11 20**.

For additional specific information about childhood cancer, contact any of the major children's hospitals and networks in your state or territory.

Chance of cure

Many children with cancer are cured of the disease. Children's bodies have great capacity for healing. Also, huge improvements have been made in the treatment of childhood cancer in the past few decades. In the 1980s, around 65% of children diagnosed with cancer were alive more than 5 years after their diagnosis. Today, around 83% of children are successfully treated and become long-term survivors.

Long-term survival (also called the outlook or prognosis) and treatment options depend on a range of factors, including:

- age of your child at diagnosis
- extent or stage of the cancer
- appearance of the cancer cells under the microscope (the shape, function and structure of the cells)
- how the cancer responds to treatment
- cancer or tumour biology, which includes
    - the patterns of the cancer cells
    - how different the cancer cells are from normal cells
    - how fast the cancer cells are growing.

Talk to your child's doctor about your child's individual disease, treatment options and outlook.

Clinical trials

Researchers are trialling new ways to diagnose and treat different types of cancer. Your child may be invited to be part of a clinical trial to test new ways of treating retinoblastoma.

New treatments have to go through very strict regulation and approval processes before they can be used in a clinical trial. Your child's doctor will explain everything about the trial and give you detailed written information. You will need to give special permission for your child to be part of the trial.

Participating in a clinical trial may or may not directly benefit your child, but the results of clinical trials today will help children with cancer in the future.

See Clinical trials and research for more information, including whether there are any clinical trials your child can join.

More information

For more information about retinoblastoma, see:

- Retinoblastoma, from the American Cancer Society
- Retinoblastoma treatment (PDQ®), from the National Cancer Institute (United States).
- Paediatric Cancer Registry